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A PRACTICAL DISCUSSION OF DISEASES AFFECTING THE BLOOD VESSELS OF THE EXTREMITIES

Knowledge of a working classification of diseases of blood vessels of the extremities, and of the differential diagnosis of these diseases, eliminates most of the difficulty in diagnosis, and therefore in decid-

ing on treatment.

Instruments of precision, for elaborate studies, are not necessary in the average case. Diseases of the peripheral arteries of the extremities fall into two large groups: (1) functional and (2) organic. Of the vasomotor or functional group of diseases, there are two main types: (1) the vasoconstricting type accompanied either by the multiple phase color reaction (Raynaud's disease) or by the one phase color reaction (acrocyanosis) dead fingers and local syncope; (2) the vasodilating type, of which ery-thromelalgia is a well known example. In the organic group, there are five main divisions: (1) arteriosclerosis with or without thrombosis, (2) thrombo-angiitis obliterans (Buerger's disease), (3) arteriovenous fistula (congenital and acquired), (4) simple thrombosis and embolism and (5)

aneurysm with or without thrombosis.

Sufficient information to allow of diagnosis usually will be obtained by the following: eliciting a history of signs and symptoms of circulatory disturbances in the extremities; examination of the hands and feet with reference to changes in temperature, trophic changes, and the presence or absence of rubor when the extremity is dependent, or of blanching when the extremity is elevated; palpation of the brachial, radial and ulnar arteries, and palpation of the femoral, popliteal, dorsalis pedis and posterior tibial arteries. The difficulty in diagnosing localized vascular disease of the extremities is in distinguishing the vasomotor or functional type from the organic type, and particularly in recognizing these lesions when they are associated. Raynaud's disease affects young women in more than 95 per cent of instances. Because it is caused y vasospasm it is completely relieved, if uncomplicated, by sympathetic ganglionectomy. This is used in severe cases. The first symptoms of the condition generally appear in winter, and consist of changes in color, usually of all of the fingers or toes. The involvement is symmetrical. The changes in color are of the so-called three phase type. The patients frequently note that the digits become white and "dead" with exposure to cold, and when warmed they become red, or often blue. These changes in color frequently are excited by emotional disturbances. The peripheral arteries of the involved extremities always pulsate normally. Hence, the pain of intermittent claudication never occurs in Raynaud's disease. Gangrene, when present in advanced cases, is limited to the cutaneous surface, in contradistinction to the mass gangrene which sometimes occurs in thrombo-angiitis obliterans.

Mild cases of Raynaud's disease usually do not require treatment. The patients should refrain from exposure to cold, and are better if they spend the winter in a warm climate.

The incidence of erythromelalgia is approximately one case in every 200 cases of peripheral vascular disease. It is the rarest of peripheral

vascular diseases.

G. E. Brown outlined four criteria for diagnosis of this disease: (1) bilateral burning pain in the extremities, (2) sharp increase of local heat in the affected parts, but redness, flushing or congestion may vary in degree, (3) production and aggrava-tion of the distress by exposure to heat and by exercise, and (4) relief by rest, cold, and elevation. Pulsations in the peripheral arteries are either normal or accentuated. Intermittent claudication and gangrene never occur. The temperature of the involved extremities is always high.

Arteriosclerosis with occlusion affects men who are more than fifty-five years of age in more than 90 per cent of cases, and since vasospasm is rarely, if ever, an element in the condition, it is not affected by sympathetic ganglionectomy. Arteriosclerosis, with or without arterial occlusion, is common and usually affects men who are more than fifty-five years of age, in contradistinction to thrombo-angiitis obliterans which usually affects young men. Pulsations of the peripheral arteries in arteriosclerotic disease of the extremities are usually diminished, and frequently pulsations are absent in the popliteal, dorsalis pedis and posterior tibial arteries. Occasionally the femoral artery is occluded, but it is rare to find a radial, ulnar, or brachial artery occluded.

Intermittent claudication is the most significant symptom in the diagnosis of occlusive arterial disease of the extremities. It is a symptom and not a disease. It manifests itself following exercise as a cramp-like pain or as a feeling of distress in the upper or lower extremities. It frequently is noted in thrombo-angiitis obliterans, and may affect the upper extremities. It rarely is observed in arteriosclerotic disease. In the lower extremities, the distress is usually confined to a single digit, to the arch of the foot, or to the calf of the leg. In the upper extremities, either a digit, the entire hand, or the forearm may be the site of distress. It is the result of an insufficient supply of arterial blood to the distal parts. It invariably comes on after exercise and is relieved by rest. Many patients can state the number of steps necessary for the production of pain, and the period of rest required for relief. More than 50 per cent of patients who have occlusive vascular disease of the extremities give histories of intermittent claudication. Since intermittent claudication never occurs in subjects with Raynaud's disease or erythromelalgia, the symptom practically means either arteriosclerosis with occlusion or thrombo-angiitis obliterans. In the presence of intermittent claudication, the brachial, radial and ulnar arteries, and the femoral, popliteal, dorsalis pedis and posterior tibial arteries, should be examined to determine whether pulsations are present or absent.

The roentgenographic demonstration of calcifica-tion in the arteries of the extremities does not necessarily mean that the patient has primary arteriosclerotic disease, for many subjects in whom

thrombo-angiitis obliterans has developed earlier in life may, in the fifth or sixth decade, show evidence of calcification of the peripheral arteries.

Thrombo-angiitis obliterans is a chronic disease of the arteries and veins which is largely confined to the extremities and occurs predominantly among men between the ages of twenty-five and fifty years. Approximately 99 per cent of the subjects affected are males. The reason for this is not clear. The disease is characterized by a chronic relapsing lesion of the vessels, and occlusion and collateral circulation struggle for supremacy. The element of time is most important in this struggle, since on this rests preservation of the part. If the intervals between relapses are short, and the time for adequate collateral circulation is brief, trophic changes and gangrene are likely to ensue. Conversely, if the intervals of time between relapses are long, collateral circulation becomes adequate, and sufficient supply of blood to the distal parts is assured. The first consideration regarding prognosis is the frequency of exacerbations of the disease. Experience in a large number of cases of thrombo-angiitis obliterans shows clearly that a high percentage of bad prognoses (regarding amputation) is not justified in this disease. Brown and I (1933) reported ten cases of thrombo-angiitis obliterans affecting women. We could find in the literature only three additional reports of cases in which the patients were women. This disease has been observed in persons of practically every nationality.

The etiology of thrombo-angiitis obliterans is as yet unknown. The excessive use of tobacco seems to aggravate the disease, but has not been identified as the causative agent. Berger reproduced the lesion in human beings by implants of segments of diseased veins. Horton and Dorsey reproduced the lesions by injection of organisms isolated from acutely inflamed veins of man and also by implantation, adjacent to the vessels of experimental animals, of segments of acutely inflamed veins of

man.

The pain of intermittent claudication is the predominating, presenting symptom in cases of thrombo-angiitis obliterans. Palpation of the brachial, radial, and ulnar arteries and of the femoral, popliteal, dorsalis pedis and posterior tibial arteries, invariably will show one or more of these arteries to be occluded. If circulatory insufficiency is present in an extremity, regardless of the nature of the occlusive process, there is rubor with dependency, and excessive pallor with eleva-

tion.

Superficial phlebitis of the migratory type is present in about 30 per cent of cases of thromboangiitis obliterans. It rarely, if ever, occurs in subjects with arteriosclerosis. Changes in color of the one phase or three phase type, are present in the involved extremities in 30 per cent of cases of thrombo-angiitis obliterans. It is for this reason that one is likely to confuse Raynaud's disease with thrombo-angiitis obliterans. Changes in color, of the three phase type, are always present in Raynaud's disease, and without these changes in color one could not make a diagnosis of Raynaud's disease; on the other hand, vasospastic disturbances of the three phase type, secondary to the occlusive vascular process, occur in 30 per cent of subjects with thrombo-angiitis obliterans. In thromboangiitis obliterans pulsations will invariably be absent in one or more of the usual palpable arteries of the extremities, whereas in Raynaud's disease pulsations in the peripheral arteries are normal.

The treatment of arteriosclerotic disease, with occlusion and thrombo-angiitis obliterans, consists first in prophylactic measures. These include avoidance of trauma and of exposure to excessive cold, the wearing of proper shoes, and particularly the proper surgical handling of all ingrown toe nails. Increasing knowledge by members of the medical profession, of the danger of operating on affected digits to which the supply of blood is already diminished perhaps will accomplish greater changes in prognosis than any other single factor. A red,

cold foot, which is invariably seen in cases of circulatory insufficiency due either to arterioscleross or thrombo-angiitis obliterans, must be distinguished from a red warm foot, which is secondary to some local infection around the nails or digital The measures of second importance are applied to increase the circulation in the extremities. Postural exercises, contrast baths, radiant heat, tissue extract and typhoid vaccine given intravenously, constitute the important medical measures. Roth and Barker have shown that intramuscular injection of pancreatic tissue extract (insulin free) given in doses of 3 c.c. enables many subjects with intermittent claudication to walk two to three times as far as before, without development of the usual distress. The exact way in which the tissue extract acts is not clear. From the surgical standpoint, in selected cases of thrombo-angiitis obliterans, sympathetic ganglionectomy gives exceptionally satisfactory results, because of the element of vasospasm which is superimposed on the occlusive vascular process. The effectiveness of this procedure, however, rests on the proper selection of cases. Two important points must be considered. First is the determination of the amount of vasoconstriction in the collateral circulation (by the fever test originated by Brown), and second is evaluation of all the additional clinical data, such as age, nationality and occupation of the patient, the rapidity of development and duration of the disease, the history of relapse, the presence or absence of trophic changes, and the degree of disability. These should be considered before deciding whether a given subject is or is not suitable for sympathectomy.

No difficulty should be experienced in the diagnosis of acquired arteriovenous fistula. There is always a history of injury, generally a puncture or a bullet wound. There is an increase in the surface temperature of the involved extremity and almost invariably bruit and thrill. The important points to remember in the diagnosis of congenital arterioven ous fistula are the increase in size and length of the involved extremity, the increase in surface tem perature, and the occasional occurrence of bruits and thrills. The diagnosis can be established by removing blood under oil from one of the regional veins and studying the oxygen content. In arteriovenous fistula, either acquired or congenital, there is a high admixture of arterial blood, either in the regional or deep veins. If one compares a sample of blood removed from one of these veins with blood removed in a similar manner from the op-posite extremity, the difference in color of the specimens usually is sufficient to establish the diagnosis. Bright red blood invariably is found in the regional veins of the involved extremity, whereas dark red blood is found in the veins of the normal extremity. Arteriography by means of thorotrast now offers additional means of visualizing arterio-

venous fistulas.

Simple thrombosis and arterial emboli in extremities are not observed as often as in the preceding groups of cases. Simple thrombosis occurs more frequently among subjects who have arteriosclerotic disease than among those of any other group. Frequently it is difficult to distinguish be tween simple thrombosis, particularly in the popliteal artery, and embolus. Patients usually complain of sudden pain in the popliteal space, with coldness and numbness of the leg, and it is often difficult to determine whether this is due to an embolus, or to simple thrombosis of the artery. In elderly people, in the absence of auricular fibrillstion or endocarditis, it is relatively safe to assume that simple thrombosis of the artery has taken place. Embolectomy in the arteriosclerotic group of cases is not a very satisfactory procedure.

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CORRECTION: Dr. William S. McCann of Rochester, New York, was the author of the July number of "Modern Concepts of Cardiovascular Disease." We wish to apologize for the error made in the spelling of his name.

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